CLAIMS

1. A transgenic mouse in contact with a suspected modulator of effects associated with congenital heart disease, wherein the genome of said mouse comprises a heterozygous disruption of the *CCN1* gene.

2. The mouse of claim 1, wherein said mouse is predisposed to atrioventricular septal defects.

3. The mouse of claim 1, wherein said mouse has atrioventricular septal defects.

4. The mouse of any one of claims 1-3, wherein said mouse is an embryo.

5. A homogeneous population of transgenic mice whose genome comprises a heterozygous disruption of the *CCN1* gene, wherein said mice are predisposed to have atrioventricular septal defects.

6. The mice of claim 5, wherein said mice have atrioventricular septal defects.

7. The mice of any one of claims 5-6, wherein said mice are embryos.

8. The mice of claim 7, wherein one or more of said mice are in contact with a suspected modulator of effects associated with congenital heart disease.

9. The mice of any one of claims 5-6, wherein one or more of said mice are in contact with a suspected modulator of effects associated with congenital heart disease.

10. A method of producing a mouse with atrioventricular septal defects, comprising:

(a) producing a transgenic mouse whose genome comprises a heterozygous disruption of the *CCN1* gene;

(b) testing the transgenic mouse for the presence of a phenotype associated with atrioventricular septal defects; and

(c) isolating a transgenic mouse that has a phenotype associated with atrioventricular septal defects.

11. A mouse produced by the method of claim 10.

12. The mouse of claim 11, wherein said mouse is an embryo.

13. A method of isolating a mouse with atrioventricular septal defects, comprising,

(a) testing a transgenic mouse whose genome comprises a heterozygous disruption of the *CCN1* gene for the presence of a phenotype associated with atrioventricular septal defects; and

(b) isolating a transgenic mouse that has a phenotype associated with atrioventricular septal defects.

14. A mouse isolated by the method of claim 13.

15. The mouse of claim 14, wherein said mouse is an embryo.

16. A method of identifying a mouse with atrioventricular septal defects, comprising testing a transgenic mouse whose genome comprises a heterozygous disruption of the *CCN1* gene for the presence of a phenotype associated with atrioventricular septal defects.

17. A mouse produced by the method of claim 16.

18. The mouse of claim 17, wherein said mouse is an embryo.

19. A method of identifying a modulator of symptoms associated with atrioventricular septal defects, comprising:

(a) contacting a transgenic mouse whose genome comprises a heterozygous disruption of the *CCN1* gene with a suspected modulator;

(b) measuring a phenotype associated with atrioventricular septal defects, whereby a modulator is identified by altering the phenotype in comparison to a control.

20. The method of claim 19, wherein said mouse is an embryo.

21. A method of identifying an animal that is predisposed to atrioventricular septal defects, comprising detecting the presence of an alteration in one or more alleles of the *CCN1* gene in a sample comprising DNA isolated from said animal.